



Case Report

Solid Pseudopapillary Neoplasm of Pancreas: Case Report of a Rare Tumor in a 52 Year Old Female

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ABSTRACT

Solid pseudopapillary neoplasm (SPN) is a clinico-pathologically distinct rare neoplasm accounting for 1-2% of all exocrine pancreatic tumours. It is usually reported in young females and has good long survival after surgical resection. In this report, we present a case of a 52 year old female with solid pseudopapillary neoplasm of pancreas.

Keywords: pseudopapillary neoplasm, pancreas, tumor

INTRODUCTION

Solid pseudopapillary neoplasm (SPN) is a clinico-pathologically distinct rare neoplasm accounting for 1-2% of all exocrine pancreatic tumors. [1] It was first described by Frantz in 1959 as "Papillary tumour of pancreas, benign or malignant". [2] Due to its rarity, its exact origin is yet to be established. Usually occurs in young females with a male: female ratio of 1:9.5. It has an indolent behaviour with an excellent 5-year survival. [3] It is primarily a benign tumor although perineural, perivascular invasion and metastasis have been reported in literature with good long survival following surgical resection. [4,5] We present a case of solid pseudopapillary neoplasm of pancreas with atypical presentation in a 52 year old female.

CASE REPORT

A 52 years old female presented to our hospital with upper abdominal pain for 1 month which was dull aching in nature and radiating to the back. On examination there was mild tenderness in epigastric region. Lab investigations included liver function test, serum amylase, serum lipase and CA 19.9 levels; all of which were within normal limits. USG abdomen was suggestive of retro-gastric mass lesion with a possibility of neoplasm of pancreas. On CT scan possible diagnosis of mucinous cystic neoplasm of pancreas involving the tail of pancreas was given. With this diagnosis distal pancreatectomy with splenectomy was performed. Intra-operatively tumour was seen to be arising from the pancreas and encasing coeliac axis, hepatic artery and gastric artery. Grossly, resected specimen of pancreas showed an encapsulated tumor with mucinous, cystic, necrotic and solid

areas. (Fig 1) On microscopic examination, section showed tumor composed of sheets, cords, trabeculae and pseudopapillae of round epithelial cells with eosinophilic cytoplasm, ovoid nuclei with finely dispersed chromatin, inconspicuous nucleoli and few with nuclear indentations along with delicate intervening vasculature. (Fig 2,3) The tumor cells were positive for vimentin, CD 56 and CD 10. (Fig 4,5,6) Based on the microscopic features and immunohistochemical (IHC) staining pattern, a diagnosis of solid pseudopapillary neoplasm of pancreas was given.

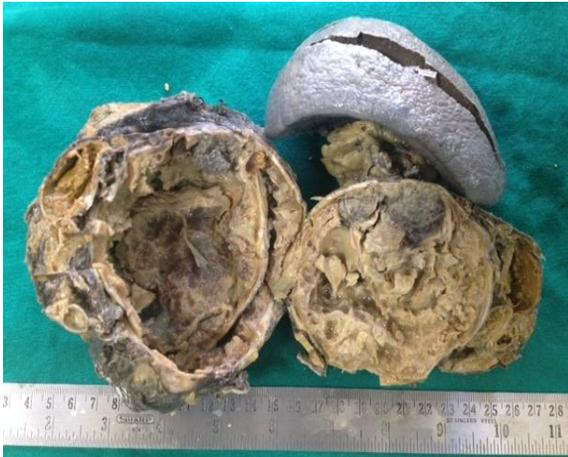


Figure 1- Encapsulated tumor of pancreas. On cut section mucinous, cystic, necrotic and solid areas noted. Also seen attached spleen.

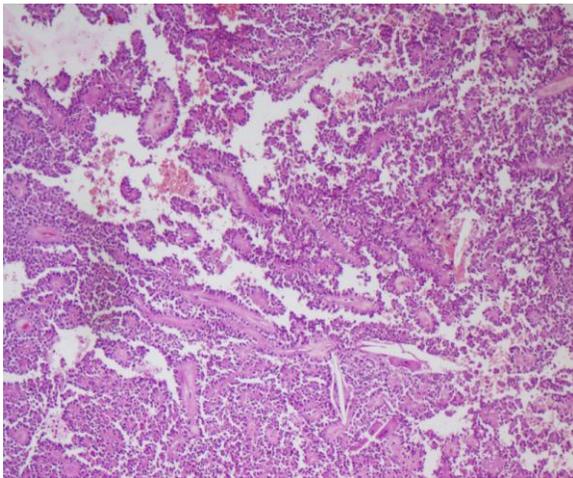


Figure 2- Tumor composed of sheets, cords, trabeculae and pseudopapillae H & E X100

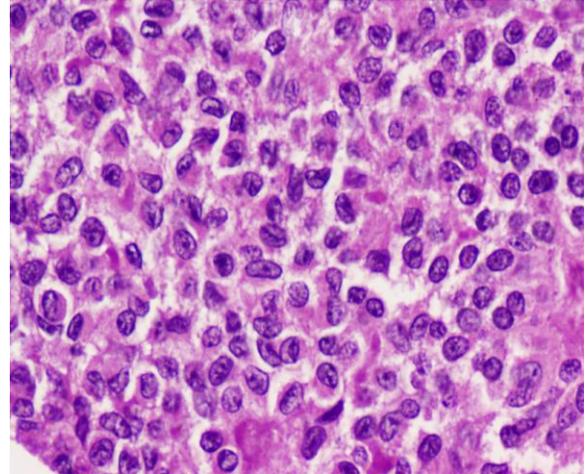


Figure 3- Tumor cells with eosinophilic cytoplasm, ovoid nuclei with finely dispersed chromatin, inconspicuous nucleoli and few with nuclear indentations H& E X400

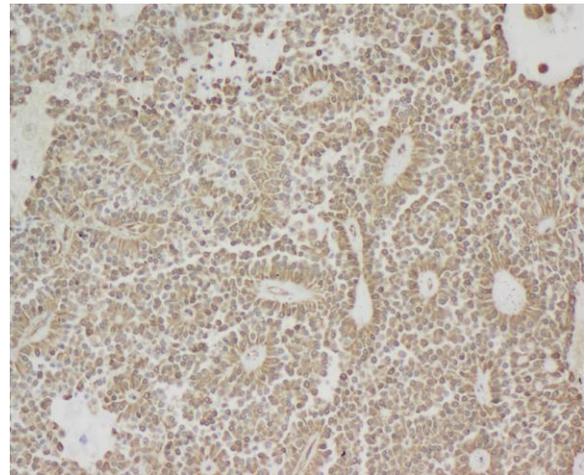


Figure 4- Tumor cells positive for Vimentin X100

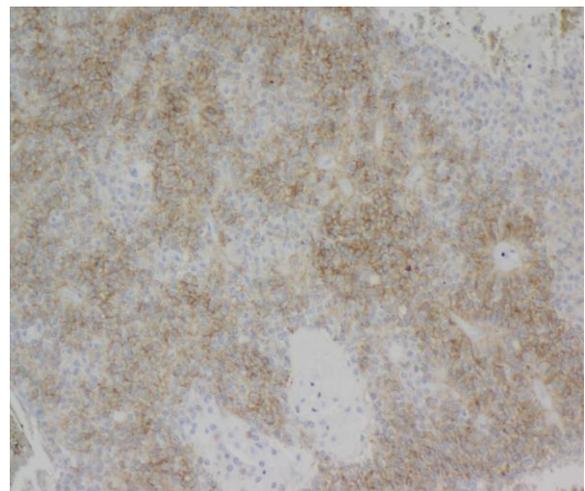


Figure 5- Tumor cells positive for CD 56 X100

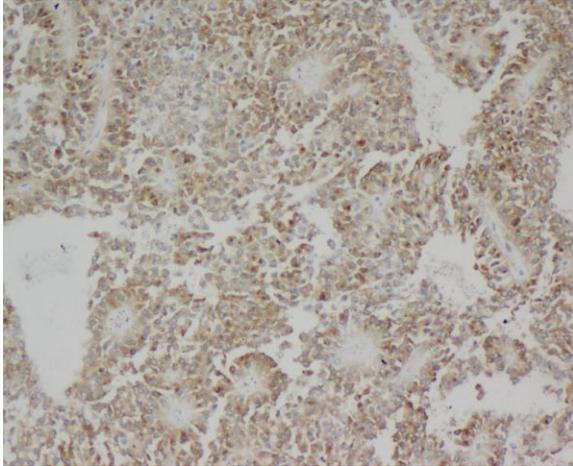


Figure 6- Tumor cells positive for CD10 X100

DISCUSSION

Originally described by Frantz, SPN had been known by multiple names such as solid and cystic tumor, solid and papillary epithelial neoplasm, papillary-cystic neoplasm, papillary cystic epithelial neoplasm, papillary-cystic tumor, and Franz tumor. [6] However, in 1996, the World Health Organization (WHO) renamed this tumor as solid pseudopapillary neoplasm. [7] According to WHO classification, SPN with invasion of adjacent structure or metastasis were called as solid pseudopapillary carcinoma. [4] It mostly occurs in young females in second to fourth decades of life. [8] However in unusual presentation it can occur in elderly individuals, children and male patients as seen in our case. Tumors in older age group and male gender are more likely to be malignant and presents with metastasis or invasion of adjacent nerves and vessels. [4] The most common site of metastasis reported are liver and omentum. [7] In our case the female was 52 years old with an encapsulated tumor encasing coeliac axis and gastric and hepatic artery but had no signs of metastasis until the last follow up. The clinical course was uneventful following surgery. Patients with SPN usually present with vague abdominal pain and in some cases with jaundice especially if

the tumor arises from the head of pancreas. However, in many cases it is an incidental finding on radiology done for other reasons. SPN can arise from any part of pancreas – head, body or tail; tail being the most common site. [6,9] On radiological examination, SPN occurs as a well circumscribed tumor with solid and cystic areas. Focal areas of calcification may be present. [3] Radiologically, it can be confused with pseudocyst pancreas, serous cystadenoma, mucin producing tumors and islet cell tumors. In children, a differential of pancreatoblastoma should also be considered. [4] On gross examination the tumor is usually well circumscribed and encapsulated with solid and cystic components along with areas of haemorrhage and necrosis. On microscopic examination, solid areas and pseudopapillary areas are seen which are composed of monomorphic cells with cytoplasmic globules. The nuclei are round to oval with fine chromatin and nuclear grooves. The papillary architecture in this tumor occurs due to cystic degeneration, thus they are not true papillae but “pseudopapillae”. Surrounding these areas foamy macrophages, cholesterol clefts and calcification may be seen. SPN stains positive for NSE (neuron specific enolase), CD 56, CD 10, vimentin, α 1-antitrypsin, α 1-antichymotrypsin and focally for synaptophysin and are negative for chromogranin, epithelial membrane antigen and cytokeratin. [10] Also nuclear and cytoplasmic overexpression of β -catenin may be observed in 95% of SPNs. [11] Based on morphology and immunohistochemical findings, SPN should be differentiated from its two close mimics, primary neuroendocrine tumor (PNT) and adenoid cystic tumor (ACT). According to Notohara et al, [10] diagnosis of SPN over PNT should be considered if cells show strong reactivity for CD56 and CD10, and focal positivity for

other neuroendocrine markers along with absence of chromogranin. Conversely, PNT shows more diffuse immunoreactivity for various neuroendocrine markers, and weak positivity for CD56. ACT are negative for CD56 and shows expression of acinar and ductal markers such as cytokeratins which helps in distinguishing it from SPN. According to Niu ZX et al [12] tumor cells of SPN express exocrine character with neuroendocrine differentiation. Hence, origin from pluripotent embryonic cells of the pancreas with multipotential differentiation was probably suggested. SPN shows expression of both endocrine (NSE and CD56) and exocrine markers (vimentin, alpha-1 antitrypsin). However there is no clear cut differentiation to any terminal pancreatic cells so the origin of tumor cells in SPN is still controversial. Kosmahl et al [13] have suggested origin of SPN from genital ridge hypothesising that surface ovarian cells could have got incorporated in pancreas during development phase when they lie in close proximity to each other. This explains similar IHC profile as ovarian tumors, positivity for progesterone receptor and predilection towards young females.

SPN is an indolent tumor with low malignant potential and excellent prognosis. Surgery is the treatment of choice even if the tumor invades the adjacent structures. [6] Complete excision of the tumor should be attempted with reconstruction of the blood vessels. [14] An excellent 5-year survival of 95% is usually seen following surgery. [5] However, a local recurrence rate of 6.2% and distant metastases of 5.6% have been reported in literature. [15] Even with recurrence and metastasis patients have good long term survival.

CONCLUSION

A diagnosis of SPN should always be considered in any young female patient presenting with a solid and partly cystic

mass of the pancreas. This tumor has a good overall prognosis following surgical resection with minimal long term morbidity. [14] It exhibits a unique IHC features with expression of CD56, CD10, and focal expression of other neuroendocrine markers. Although, the clinical signs and symptoms are relatively nonspecific, imaging with histopathology and IHC can help separate these tumors from the other malignant pancreatic tumors.

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